280* Validation of the MST-25: an extension of the modified shuttle test (MST)

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The Modified Shuttle Test (MST) is an externally paced, incremental, field exercise test with 15 levels. It has been shown to be a reliable, repeatable and sensitive measure of exercise capacity in adults with cystic fibrosis (CF). In our clinic, however, 6% of patients exceed the 15th level of the test. Furthermore, 31% of age-matched healthy adults exceeded the 15th level. This limits the utility of the MST as a measure of exercise capacity in studies comparing participants with CF to healthy control participants. The purpose of this study was to assess the reliability of the MST with 25 levels (MST-25).
Method: The MST was extended by 10 levels, with the number of shuttles increased by one at each additional level. 15 participants (10 healthy, 5 CF) performed the MST-25 on two occasions within one week. All participants were clinically stable (<10% variation in FEV1 from the best during the previous 6 months). All participants wore the SenseWear Pro3 Armband (BodyMedia, USA) – a physical activity monitor that integrates accelerometry and physiological sensors to estimate energy expenditure. Total step count, Borg Dyspnoea Score and heart rate were recorded at the end of the test.
Results: There was a significant and strong correlation between the two MST-25 test days for distance completed (Pearson's r = 0.99), SpO2 at peak (r = 0.99), energy expenditure estimated by the SenseWear Pro3 Armband (r = 0.97), and total step count (r = 0.97). Correlations were less strong for dyspnoea (r = 0.76) and heart rate (r = 0.78) at peak exercise.
Discussion: The MST-25 shows good test re-test reliability and can be used for patients and healthy controls whose peak exceeds the MST. We are now investigating the sensitivity of the MST-25 to clinical change.

281* Mathematical modelling of oxygen uptake during recovery from exercise

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Objectives: Peak oxygen uptake (VO_{2peak}) derived through maximal cardiopulmonary exercise testing (CPXT) is impaired in children with Cystic fibrosis (CF), and is a measure of disease severity. Our aims were to compare the profile of oxygen uptake (VO_2) during recovery following CPXT in children with CF and controls.
Methods: 19 children with CF and 19 controls matched for age, sex and maturity performed CPXT on a cycle ergometer to exhaustion. Pulmonary gas was measured continuously by an on-line metabolic gas analyser using a 10 s moving average, during and for 10 min after CPXT. Following CPXT the fast component of VO_2 falls in an exponential manner that can be mathematically modelled, using a mono-exponential model including a delay term, with the formula; $VO_2(t) = A\{1-\exp[-(t-TD)/\tau]\}$ where; $VO_2(t)$ is VO_2 at a given time point; A is the amplitude of the fast component from VO_{2peak}; D is the delay term; τ is the time constant, the time taken to reach 63% of the amplitude of the fast component of VO_2 recovery. Disease severity in the children with CF was quantified by the Shwachman score.
Results: VO_{2peak} was reduced in children with CF compared to controls (35±8 versus 44±12 $mL \cdot kg^{-1} \cdot min^{-1}$, $P < 0.01$). Furthermore, the τ was significantly longer in children with CF compared to controls (44.9±9.8 versus 38.5±6.0 s, $P < 0.05$), and related to disease severity ($r = -0.75$, $P < 0.01$). The τ was not significantly correlated with VO_{2peak} in either the children with CF or controls. Therefore, the τ can not be reliably predicted by VO_{2peak}.
Conclusions: The pattern of decline in VO_2 during recovery after CPXT differs between CF and controls and is related to disease severity. Recovery measures may enhance our objective assessment of exercise impairment in CF.

282* Desaturation on 3-minute step test is associated with impaired outcomes at 12 months in adults with cystic fibrosis

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The three-minute step test (3MST) is a simple test of exercise capacity which is commonly used in patients with CF, however there are no data regarding its prognostic value. This study aims to determine whether performance on the 3MST is associated with long-term clinical outcomes in adults with CF.
Methods: Clinically stable patients were recruited from the CF outpatient clinic. The 3MST was conducted following a standardised protocol using a 15cm high step, external pacing at 30steps/minute and continuous monitoring of SpO_2, heart rate and perceived exertion on the Borg scale. Relationships between baseline performance on the 3MST and clinical outcomes at 12 months (change in % predicted FEV_1 and inpatient hospital days) were assessed.
Results: Data were available for 48 (30 males) of mean age 30 (standard deviation 8) years, FEV_1 64 (25) % predicted and BMI 23 (5) $kg.m^{-2}$. Participants who desaturated to less than 90% on 3MST (n = 12) had a larger number of hospital days over the following 12 months than those who did not (median 17 days vs 2 days, p = 0.007). Those who desaturated also had and a greater decline in FEV_1% predicted (mean difference 4.9%, 95% confidence interval 0.5–9.3%). Heart rate and symptom scores on 3MST were not associated with clinical outcome at 12 months.
Conclusions: Performance on the 3MST may be associated longer-term pulmonary deterioration and increased hospital admission days. Larger samples are required to confirm the prognostic value of the 3MST in adults with CF.

283 Reference values for maximal aerobic exercise capacity in children with cystic fibrosis

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Introduction: In patients with cystic fibrosis (CF) exercise capacity is related to disease severity, nutritional status, lung function, quality of life, morbidity and mortality. Reference values of maximal oxygen uptake (VO2max) of healthy children are available however these are unsuitable for CF children as height-for-age and nutritional status are regularly reduced and onset of puberty is frequently delayed. CF specific reference values will give better insights in disease-related normal deterioration of exercise capacity. Therefore, the aim of the study is to create disease-specific reference values for VO2max in relation to age, sex, anthropometric values and lung function in CF children.
Methods: Data were acquired from a database (1998–2008). VO2max was measured during CPET. Anthropometric variables are expressed as weight and height and lung function as the forced expiratory volume in 1 second (FEV1). Data were analyzed with the GAMLSS method.
Results: 1109 CPET results were used of 277 CF children (146 boys; 131 girls) with a mean (SD) age of 11.84 (3.63) years, mean (SD) weight of 39.19 (14.64) kg and mean (SD) length of 147.80 (20.07) cm. FEV1 of predicted ranged from 21 to 151 with a mean (SD) of 89 (20) %. VO2max ranged from 0.30 to 3.80 with a mean (SD) of 1.66 (0.61) $l \cdot min^{-1}$. Final reference equations will be presented.
Conclusion: CF-specific reference equations allow clinicians to compare exercise capacity of individual patients with those of their peers with CF. This might improve generalization of future CF clinical trials by setting entry criteria that are equitable across sex and age ranges. The new reference equations may serve as a useful adjunct to conventional ones for healthy children.